Editorial Commentary to JTCVS-17-1828R1 Feature Expert Opinion "Ask not what your Fontan can do for you, ask what you can do for your Fontan!"

The Fontan circulation: time for a moonshot?

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Central message (174/200 characters + spaces):

Fontan repair is palliative. With increasing late survival, we must now learn how to manage our success. Late Fontan problems are intractable. New therapies can prevent them.

This article reminds us of the fate of the Fontan patient, and our obligation to provide the best possible care over their lifetime [1]. It outlines the potential benefits of improving, in so far as possible, the known factors that adversely affect hemodynamics. Efforts to reduce these risk factors should be pursued when reasonable to do so. The risks of multiple reoperations will, however, need to be balanced against the potential, but as yet unproven, benefits.

At the same time, the article reminds us that Fontan repair is palliative [2-4]. After 5 decades, the long-term fate of the Fontan circulation is now clearly framed as an intractable problem that culminates in a host of secondary diseases [5]. After all, the absence of a subpulmonary ventricle is physiologically profound. Medical therapies do not provide clear solutions. Late surgical optimization of correctible factors is important, but cannot overcome the non-correctible factors.

Mechanical circulatory support (MCS) has, up to now, generally focused on the application of systemic support in end-stage Fontan disease using existing devices. However, this is highly problematic. Existing MCS technology is better suited to ventricular failure in Fontan, rather than Fontan failure secondary to lack of a subpulmonary ventricle. In the setting of preserved systolic function (>70% at late follow-up [6]), systemic MCS is superfluous; in fact, it may further congest the right-sided circulation where the circulatory bottleneck exists, and exacerbate Fontan disease. Transplantation is not a comprehensive solution, and represents end-stage therapy.

Current therapies are incremental and aimed at optimizing palliation; they are not capable of resolving the underlying physiologic deficit (i.e. cure). So, where do we go from here?

A fundamentally different approach is possible with curative potential for our single ventricle patients. Biventricular equivalency can be maintained in the single ventricle circulation with the use of cavopulmonary assist, preempting Fontan disease [7]. A highly specialized, relatively innocuous, low-pressure (~6 mmHg) cavopulmonary assist device will reverse the Fontan paradox by reducing systemic venous pressure (~6 mmHg) and improving preload (~2 mmHg), thereby improving cardiac output. A durable subpulmonary power source will permanently normalize the circulation.

Rather than applying MCS for end-stage salvage, we need to apply it preemptively for biventricular health maintenance. Rather than Fontan perpetuation, we need Fontan reversal.

To paraphrase the venerable words of John F Kennedy, I believe that this [field] should commit itself to achieving the goal, within a decade, of [reversing the Fontan and emulating biventricular circulatory health in single ventricle patients]. A Fontan-specific device is necessary; existing technology will not scale [8]. Devices that can safely and reliably achieve this vision are in development and within reach.

The treatment of single functional ventricle has not significantly changed for decades, succeeding only in prolonging an inherently inefficient system. To change long-

term outcomes, it is time to replace what is missing. We can solve the unimaginable and shift the paradigm for single ventricle care to one based on biventricular health. Our single ventricle patients stand to benefit greatly.

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